

**COMMONWEALTH OF AUSTRALIA**  
**Copyright Regulation**

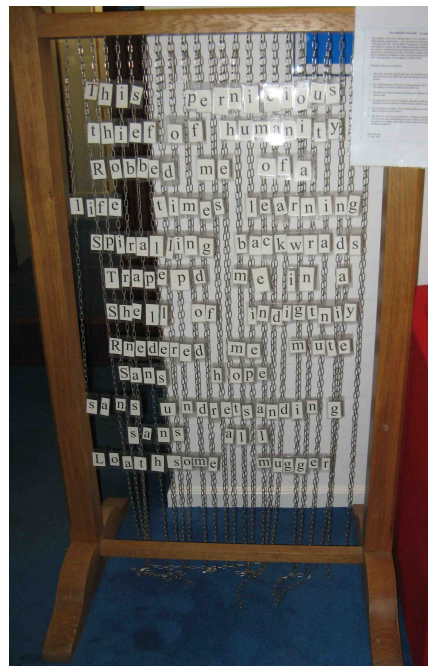
**WARNING**

This material has been reproduced and communicated to you  
 by or on behalf of the University of Sydney pursuant to Part  
 VB of the *Copyright Act* 1968 (**the Act**).

The material in this communication may be subject to  
 copyright under the Act. Any further reproduction or  
 communication of this material by you may be the subject  
 of copyright protection under the Act.

Do not remove this notice

This pernicious  
 thief of humanity  
 robbed me of a  
 life times learning  
 Spiralling backwrads  
 Trapepd me in a  
 Shell of indigtniy  
 Rnedered me mute  
 Sans hope  
 sans undretsanding  
 sans all  
 Loathsome mugger



# Educational Workshop: Assessment and Management of Progressive Aphasia in the Frontotemporal Dementias

Karen Croot

School of Psychology  
University of Sydney

Cathleen Taylor

PPA and Related Disorders Clinic  
War Memorial Hospital



Please do not photograph/film the videos of people with progressive aphasia. Some of them have consented to their video only being shown in this setting.

Please do not photograph or film the creative/art works reproduced in this presentation.

## About us, and our interest in PPA

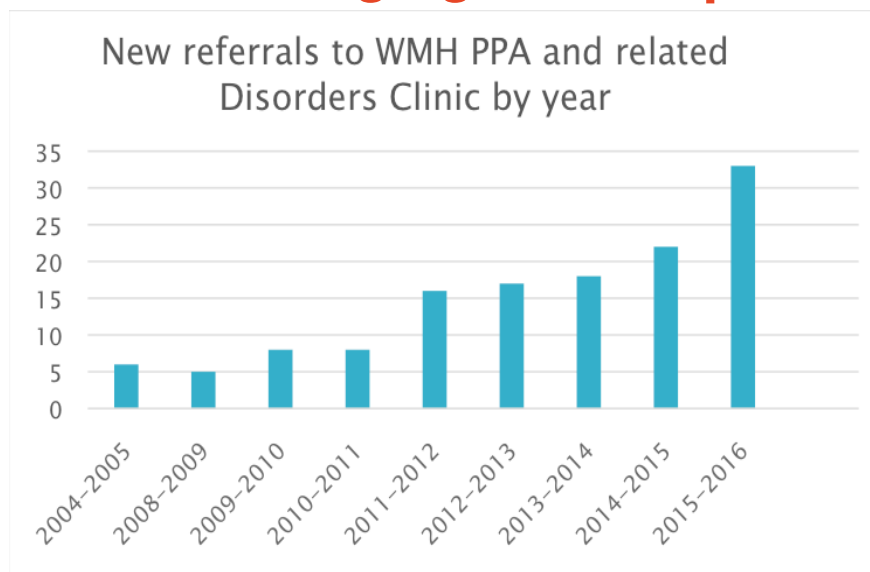
### **Karen:** Met Mr and Mrs N during PhD in England, 1993

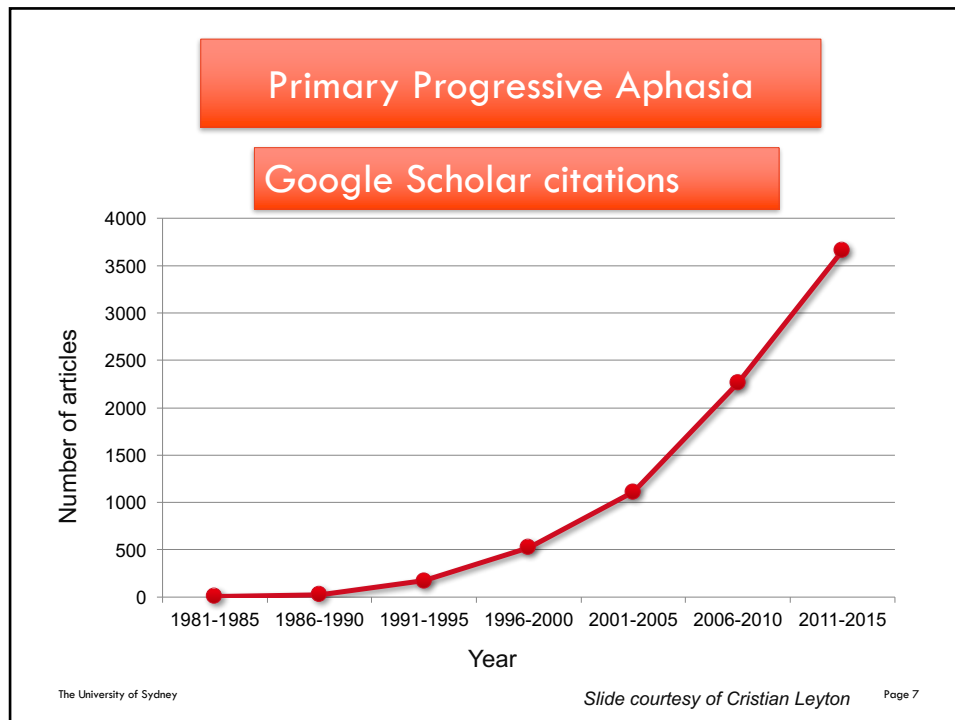
- Mr N: nfvPPA, highly communicative with writing, gesture and demonstration/enactment
- Mrs N: the stigma of “dementia” limited their social participation and made them vulnerable to depression, anxiety and relationship tension
- Mr and Mrs N needed education and supports to understand and maximise strengths and increase social participation

**Cathy:** Senior Speech Pathologist with responsibility for the PPA caseload at War Memorial Hospital

- **increasing referrals from the early 2000s**, with cases of possible PPA referred directly. Has seen >150 cases.

## PPA: An emerging area of practice





## Survey of Speech Pathology services for PPA in NSW, Australia

- Increase in referrals to War Memorial Hospital Sydney from late 1990s
- Survey of 34 speech pathology services
  - with adult neurological caseload
  - rural and metropolitan NSW
  - between June 2004 and July 2005
- Summary of responses
  - People with PPA under-referred
  - Speech pathologists felt ill-equipped
  - Overall lack of service



**(Taylor et al., 2009)**



## About you: Workshop participants

- **29 Countries:** Europe, North & South America, Middle East, Asia, Pacific region
- **Professions:** speech pathologists, neurologists, psychiatrists, geriatricians, cognitive scientists, neuropsychologists

## About a person with PPA: Introducing ANT

- Female, single, no children or siblings, lives alone
- ~ 69 years old when first noticed symptoms
- Supportive network of friends
- Power of Attorney lives ~500km away

### Occupation

- Primary school teacher for 36 years
- Retired as Deputy Principal ~65 years of age

### Family history

- First cousin with dementia

## About a person with PPA: Introducing ANT

### Summary of Case History

- 2010** Gradual difficulty word finding, expressing herself fluently, referred to speech pathologist
- 2011** Referred to geriatrician MMSE 29/30  
Neuropsychological assessment, diagnosis of possible PPA  
Brain MRI: mild-to-moderate small vessel vascular disease
- 2012** Mild decline on neuropsychological review
- 2013** Brain MRI: subtle left peri-insular atrophy with additional bilateral involvement of mesial temporal lobes
- 2014** PiB-PET scan: salient bifrontal hypometabolism, more extended in the left than the right hemisphere, and mild left hypometabolism in parietal and temporal regions.  
Non-AD pathology

This single woman continues to live independently at home 6 years after initial symptoms of PPA

## What people with PPA and their relatives and friends say in Munich

### – Individuals with PPA experience

- a high degree of psychological strain in everyday life
- social isolation

### – They hope for

*Elisabeth Wlasich & Gisela Stenglein-Krapf  
Neuropsychologists, Klinikum Grosshadern*

- a cure
- improvement in their symptoms
- Improved coping in everyday life
- opportunity to be active participants in their own care

### – They ask about

- training programs and materials they can use independently to practise language activities
- outpatient (ambulant) rehabilitation services
- support groups

13

## Overview of workshop

### – Philosophy/framework for intervention in PPA

### – Assessment

- Differential diagnosis of PPA and diagnosis of PPA variant
- Impact on everyday and functional communication
- Review assessment: Change over time
- Discharge planning and capacity assessment
- Assessment of contextual factors

### – Intervention and management

- Feedback as the 1<sup>st</sup> intervention
- Pharmacological interventions and brain health
- Speech and language abilities
- Communication activities and life participation

**Katie Brandt & Megan Quimby, MGH**

**“Harmonizing Together”**

- Personal and environmental contextual factors

The University of Sydney

Page 14

## Philosophy/framework for intervention in PPA



### Intervention *IS* indicated in neurodegenerative disease

#### Our experience

- Is it **appropriate** to intervene given trajectory of decline?, Contrasts acute-onset aphasia services to promote recovery
- Similar views challenged in Hopper (2003) **“They’re just going to get worse anyway”**: perspectives on rehabilitation for nursing home residents with dementia. *Journal of communication disorders*, 36(5), 345-359.

#### Growing appreciation of

- social and economic costs of dementia with increasing incidence
- the value in providing speech pathology and other services for dementia and PPA

## Intervention *IS* indicated in neurodegenerative disease

“Speech Pathologists need to rethink, redefine and reclaim the spectrum of clinical services that they are qualified to provide to dementia patients.”

(Mahendra & Arkin, 2003, p. 417)

The University of Sydney

Page 17

## Intervention *IS* indicated in neurodegenerative disease

### USA

- **PPA 25<sup>th</sup> Annual Research Symposium at 2015 Convention**
- Bourgeois et al. (2016). **Will You Still Need Me When I'm 64, or 84, or 104?** The Importance of Speech-Language Pathologists in Promoting the Quality of Life of Aging Adults in the United States into the Future. *Seminars in Speech and Language*, 37, 185-200.

### United Kingdom

- **RCSLT advocacy and resources**

### Australia

- **Speech Pathology Australia National Speaking Tour 2016**  
Dr Jade Cartwright: “Dementia is our Business: Speech Pathology Services for People Living with Dementia across the Continuum of Care”
- **NSW Health launching “Allies in Dementia Care”** guides for families, carers and health professionals highlighting role of speech pathologist in facilitating communication in dementia

### Munich

- Centre for Advanced Study at Ludwigs Maximillians Universität  
**Schwerpunkt: Dementia in an Ageing Society, 2011-2013**

## Speech, language and communication intervention can:

- Increase quality of life for person with PPA and significant others during the finite period of decline
- Improve communication in specific relationships
- Support instrumental end-of-life activities and decisions (guardianship, care plans, accommodation, financial arrangements)
- Support “important conversations”
- Facilitate ongoing social participation and reduce isolation (family, friendships, community)
- Contribute to emotional well-being

The University of Sydney

Page 19

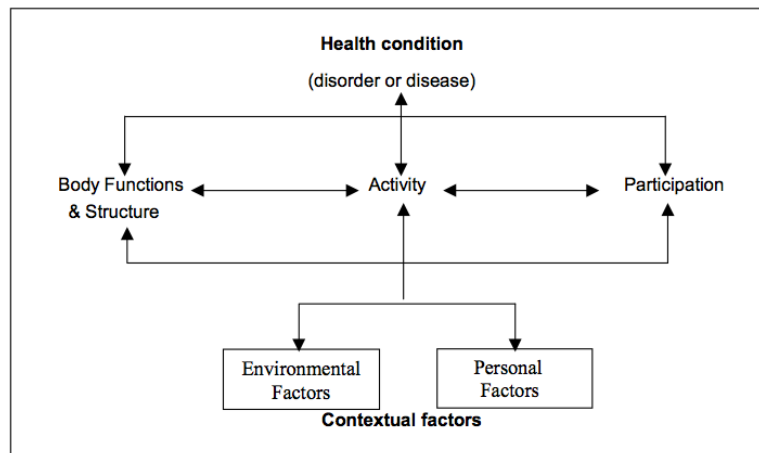
## Clinical impression of pw PPA

- Rarely deny deficits (McNeil & Duffy, 2001)
- Frequently initiate the search for “an answer”
- Do not typically complain of memory loss (except for words)
- Often no personality change, but may report frustration and/ or depression. Behavioural rigidity, lack of judgement, change in eating habits in semantic dementia.
- Drive, keep house, go to yoga, pick up the kids from school, go on overseas trips.

➔ **Aim is to maximise communication, manage other symptoms and maintain quality of life for as long as possible**

## Framework for assessment and intervention

WHO International Classification of Functioning, Disability and Health



ICF Training Beginner's Guide, available at  
<http://www.who.int/classifications/icf/icfaptraining/en/index.html>

Page 21

## Goal-setting for intervention

- Collaborative with person with PPA and significant other: relational-centred care (Morhardt et al., 2016)
- Education about disease progression
  - Realistic expectation of outcome of intervention
  - Proactive: Implementing goals in anticipation of decline (Rogers et al., 2000)
  - Expect goals and interventions to change over time
  - Increased risk of suicide soon after diagnosis: education and intervention informed by assessment of depression and suicide risk
  - Acknowledge the devastating diagnosis

The University of Sydney

Page 22

## Assessment

Differential diagnosis of PPA

Diagnosis of PPA variant

Impact on everyday and functional communication

Review assessment: Change over time

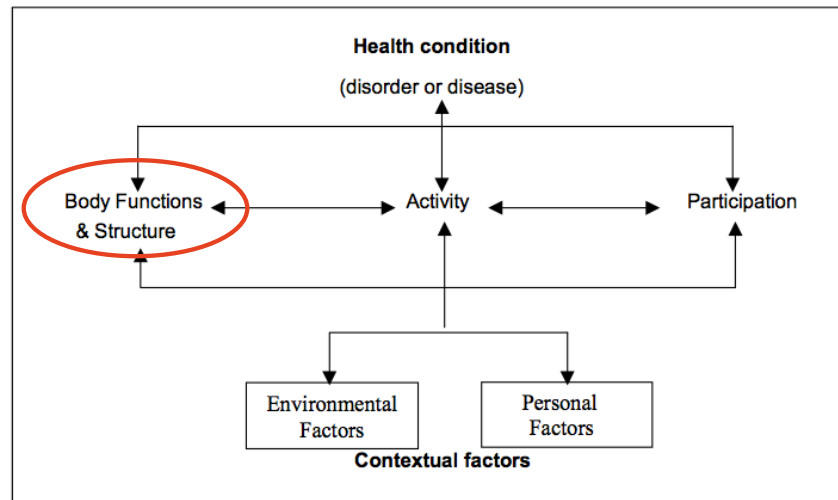
Discharge planning and capacity assessment



## Assessment for differential diagnosis of PPA



## Assessment for differential diagnosis



The University of Sydney

Page 25

## Diagnosing PPA as a syndrome caused by a disease

- **PPA is a clinical syndrome:** a collection of symptoms that reflect damage to parts of the brain due to a small number of neurodegenerative diseases
  - Diseases on the FTD spectrum
  - Alzheimer's Disease
- Initially the disease affects parts of the brain involved in language, but as the disease spreads, more abilities can be compromised. **Which ones depends on where the disease spreads.**
- People with other syndromes on the FTD spectrum may develop speech and language symptoms with progression
- Some patterns but can't definitely predict rate of decline or timing/pattern of additional symptoms for each individual. Current work by Argye Hillis and colleagues to refine prognosis on the basis of imaging.

The University of Sydney

Page 26

## PPA: Basic diagnostic criteria (Gorno-Tempini et al., 2011)

- Clinical syndrome
- NOT pathology

The University of Sydney

**Table 1** Inclusion and exclusion criteria for the diagnosis of PPA: Based on criteria by Mesulam<sup>32</sup>

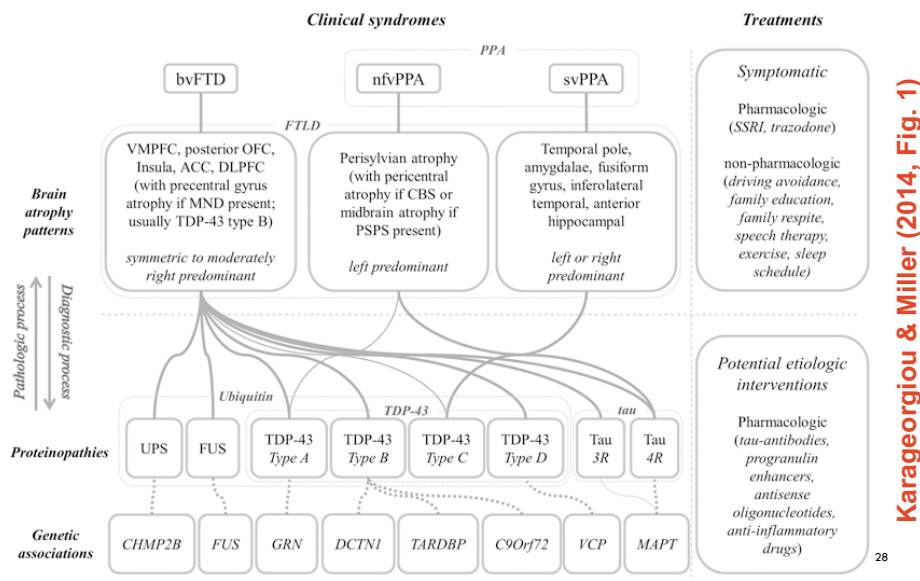
Inclusion: criteria 1-3 must be answered positively

1. Most prominent clinical feature is difficulty with language
2. These deficits are the principal cause of impaired daily living activities
3. Aphasia should be the most prominent deficit at symptom onset and for the initial phases of the disease

Exclusion: criteria 1-4 must be answered negatively for a PPA diagnosis

1. Pattern of deficits is better accounted for by other nondegenerative nervous system or medical disorders
2. Cognitive disturbance is better accounted for by a psychiatric diagnosis
3. Prominent initial episodic memory, visual memory, and visuo-perceptual impairments
4. Prominent, initial behavioral disturbance

## Relationships between clinical syndromes, brain atrophy, neuropathology and genetic mutations



## Diagnosis: Introducing TOC

- 58 year old female
- Tertiary educated businesswoman
- **October 2011** noticed changes to motor speech and subtle word finding problems
- **June 2012** sought diagnosis and contacted WMH for specialist services
- **August 2012** Discussing experience of diagnosis by exclusion: she wants to be informed, confirm what's going to happen
- In other conversations we see partner avoiding talk of prognosis: 6/7 partners (n=10) appeared to be unable to engage when the topic was prognosis/future (Taylor et al., 2015 and in prep.)

## Differential diagnosis of PPA/language variant FTD

- **Alzheimer's disease** (especially in relation to syndrome of logopenic progressive aphasia, typically associated with AD pathology): Assess immediate and delayed verbal and visual memory to determine whether person shows rapid forgetting and include a "memory profile" in report (Dr Nicole Kochan, NPI POWH Sydney). PET-PiB imaging.
- **Vascular Cognitive impairment** (may be co-morbid): assess vascular risk factors, any step-wise cognitive deterioration, attention/working memory and processing speed as well as memory testing as for AD
- **Other neurodegenerative disorder**: Assess non-language cognitive impairment, executive function, limb apraxia, visuo-spatial function, social cognition, behaviour and personality, face recognition – other syndromes on FTD spectrum, Dementia with Lewy Bodies
- **Mood and/or anxiety disorders, other psychiatric disorder**: Assess history and recent change.

### Resources:

- Clinical interview
- ACE-III Cognitive Examination III (Hsieh et al., 2013)
- Neuropsychological assessment battery

## Responses to diagnosis

- Years between first awareness and diagnosis carry their own stress (approx. 4 years, Le Rhun et al., 2005)
- Can be a relief for people with PPA and families: knowing person has a known syndrome can help with understanding and feeling understood about frustrating, “odd” or “embarrassing” behaviour. Consider providing picture of brain scan to make “invisible” disease more visible.
- Some pwPPA choose not to disclose because of ongoing social stigma
- Can provide impetus for important conversations, planning for future, including power of attorney, guardianship, future health care plans, living wills
- There may be profound adjustment and grief, increased suicidality
- There may be increased resolve: **“The enemy now has a name and I can fight that enemy.”** (Anthony, pwPPA, 2010)
- There may be different responses from pwPPA and family member

The University of Sydney

Page 31

## Diagnosis: Introducing Boyd and Shirley

Up until this time, Boyd didn't let me go in with him when he went to see the doctor. I had become very worried and distressed about his problems, because I only knew what Boyd told me. I had to force the issue and insist on being with him the next time he saw the doctor. I said to the doctor that I wasn't happy with what was happening with Boyd's condition and asked if he could refer Boyd to a specialist, to hopefully find out what was causing his speech problems, so he did.

Boyd finally got an appointment to see a neurologist in January 2009. By the end of the consultation, the neurologist said he was 90% sure that Boyd had a very rare brain disease called PPA. Three months later, it was confirmed after different tests had been done, including an MRI scan.

We had never heard of anything like this before and neither had our local doctor. It is a very rare brain disease and very difficult to diagnose. The neurologist told us that there wasn't any medication he could give Boyd for it. There was no cure. The brain was shrinking and the condition would gradually get worse. He said that it

Rutherford, 2009, p. 17

The University of Sydney

Page 32

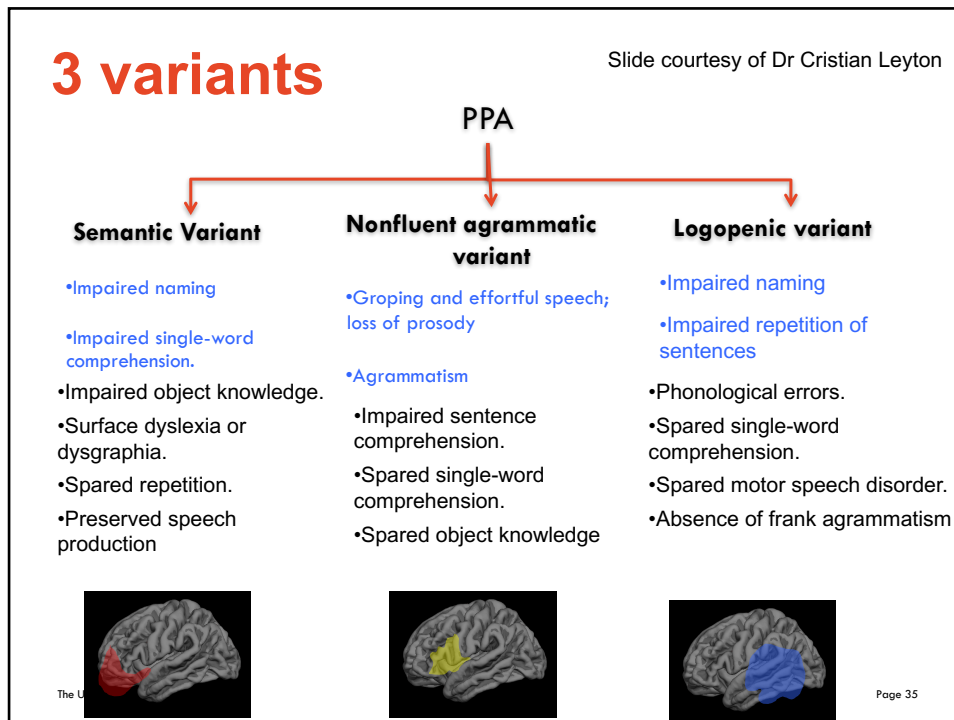
## The Sydney experience

- FRONTIER/FOREFRONT group (Hodges, Piguet, Mioshi, Hornberger) established at University of NSW in the 2000s, moving to the University of Sydney 2017
- Exponential increase in research and clinical and public education about FTD by this group in Sydney and colleagues in Melbourne and throughout Australia
- Time to diagnosis has decreased
- Most frequent referral question from GPs and consultant medical practitioners is now: **Is it PPA?**

## Assessment for diagnosis of PPA variant

## 3 variants

Slide courtesy of Dr Cristian Leyton



## Epidemiology (Karageorgiou & Miller, 2014)

**Table 1** Epidemiology of frontotemporal lobar degeneration (FTLD)<sup>19–22</sup>

Clinical syndrome	Percentage of FTLD cases	Range of male percentage	Mean age of onset (range) <sup>a</sup>	Life expectancy in years from symptom onset (from diagnosis) <sup>b</sup>
bvFTD	54–69	53–70	58 (47–82)	with MND 6 (1) without MND 9 (5)
nfvpPPA	14–35	14–63	63 (42–79)	9 (4)
r-svPPA	6–10	44–80	62 (52–85)	12 (5)
l-svPPA	9–12	52–80	59 (52–80)	12 (5)

<sup>a</sup>No statistical difference.

<sup>b</sup>Significantly shorter life expectancy only for bvFTD-MND cases.

**Table 4** Criteria for the diagnosis of bvFTD, nvfPPA, and svPPA<sup>1,3,4</sup>**Karageorgiou & Miller (2014)**

Syndrome	Possible/clinical diagnosis	Probable/imaging supported diagnosis <sup>b</sup>	Definite/pathologically or genetically proven diagnosis	Exclusionary criteria
nvfPPA <sup>d</sup>	At least one of the following: • Agrammatism • Effortful, halting speech with inconsistent sound errors (AOS) At least two of the following: • Impaired comprehension of syntactically complex sentences • Spared single-word comprehension • Spared object knowledge	All of the following: • Meets possible/clinical criteria • Imaging consistent with nvfPPA (left posterior frontoinsula)	All of the following: • Meets possible OR probable criteria • Histopathological evidence of specific pathology <sup>c</sup> and/or presence of known pathogenic mutation	• Deficits are not better explained by alternative diagnosis (nondegenerative, or psychiatric) • Prominent initial deficits are not memory, visuospatial, or behavioral
svPPA <sup>d</sup>	All of the following: • Impaired confrontation naming • Impaired single-word comprehension At least 3 of the following: • Impaired object knowledge • Surface dyslexia or dysgraphia • Spared repetition • Spared grammar and motor speech production	All of the following: • Meets possible/clinical criteria • Imaging consistent with svPPA (anterior temporal lobe)	All of the following: • Meets possible OR probable criteria • Histopathological evidence of specific pathology <sup>c</sup> and/or presence of known pathogenic mutation	• Deficits are not better explained by alternative diagnosis (nondegenerative, or psychiatric) • Prominent initial deficits are not memory, visuospatial, or behavioral

Abbreviations: AOS, apraxia of speech; CDR, Clinical Dementia Rating Scale; FAQ, Functional Activities Questionnaire; FTD, frontotemporal lobar degeneration; PET, positron emission tomography; SPECT, single-photon emission computed tomography.

<sup>a</sup>Approximately within the first 3 years from symptom onset.

<sup>b</sup>Imaging refers to structural magnetic resonance imaging atrophy, PET hypometabolism, or SPECT hypoperfusion.

<sup>c</sup>Specific pathology in 2011 PPA (primary progressive aphasia) criteria may be tau, TDP-43, Alzheimer disease, or other proteinopathy.

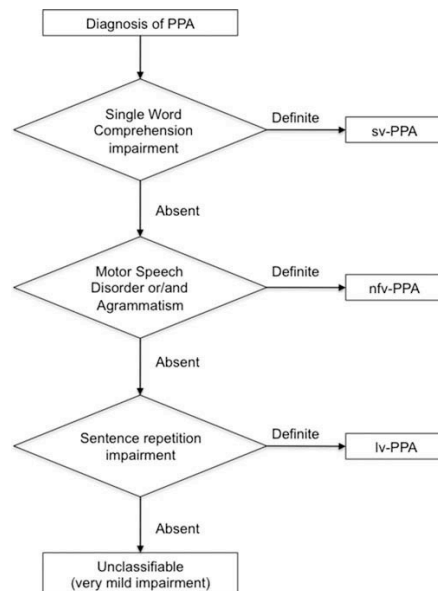
<sup>d</sup>Both nvfPPA and svPPA must satisfy PPA criteria by Mesulam<sup>1</sup> with language impairment being the most prominent, disabling, and earliest symptom.

Page 37

## And the rest... (Sajjadi et al., 2012)

- Estimates of approx 1/6 cases hard to fit into variants
  - various "non-fluent" symptoms
  - early difficulties selective for reading and/or spelling
  - late in disease progression
- Don't be afraid to say don't know diagnosis yet
- Report the clinical features and assessment results that support possible differential diagnoses
- May require review assessment (1 year, earlier if person notes significant change) to accurately diagnose PPA variant

## Assessment to classify variant



**Leyton & Hodges (2014)**

The University of Sydney

## Progressive Aphasia Language Scale (PALS) (Leyton et al., 2011)

### 15 minutes of informal spontaneous conversation

1. Observe **features of motor speech disorder** (distortions, effortful speech, abnormal prosody)
2. Observe **phonological errors** (substitution or addition of well-formed phonemic segments, deletion of segments or syllables)
3. Observe **agrammatic features** (grammatical omissions, morphological errors, predominance of short, simple sentence structures)

### Score on a 4-point scale

0 = None, 1 = mild or doubtful, 2 = definite, 3 = severe

The University of Sydney

Page 40



## Progressive Aphasia Language Scale (PALS) (Leyton et al., 2011)

### Rate performance on language tasks

1. **Naming:** 10 animal models, 10 household objects

2. **Single word repetition**

Words of increasing length and complexity (banana → chrysanthemum)

3. **Single word comprehension**

Ask about animals in naming task and definition of words in repetition task. Which one is indigenous? What sort of a thing is a catastrophe?

4. **Sentence repetition**

Repeat sentences e.g. **The lady delivered some delicious gingerbread.**

### Score on a 4-point scale:

0 = None, 1 = mild or doubtful, 2 = definite, 3 = severe

The University of Sydney

Page 41

## Additional resources for speech and language assessment

- Clinical Interview: self- and other-report of symptoms, observation of speech, language and communication strengths and weaknesses
- PALS
- FRONTIER/FOREFRONT tests e.g. SYDBAT (Naming, Repetition, Word Comprehension, Semantic Association), Repeat and Point
- Subtests from aphasia batteries (WAB, BDAE, CAT, PALPA, AAT) for hypothesis testing
- Difficult naming test e.g. GNT (sensitive in early stage)
- Syntax assessments NAT, NAVI, SECT, MAST
- Picture description (connected speech)
- Apraxia of Speech Rating Scale (ASRS) (Strand et al., 2014), ABA2 Apraxia Battery for Adults (Dabul, 2000)
- Orobuccal apraxia assessment: indicative of nvPPA and flags the need to monitor swallowing over time

The University of Sydney

Page 42

### German assessments: Language assessments used by Prof Adrian Danek's group (Munich)

- Subtests of the Aachener Aphasie Test: spontaneous speech, Token Test, Reading & Writing
- Picture description task (e.g. Cookie theft picture)
- German Repeat and Point test (unpublished)
- Picture naming: 15-item Boston Naming test
- Word fluency tasks (CERAD battery)
- Bogenhausener Semantik Untersuchung (BOSU)

The University of Sydney

Page 43



### German assessments: Materials produced by the EKN (Munich)

- BoDYS – Dysarthria Scale
- Acoustic and intelligibility analyses of dysarthria
- HWL – Word list for AOS diagnosis
- Non-verbal Semantic test

[http://www.ekn.phonetik.uni-muenchen.de/diagnostik\\_therapie/index.html](http://www.ekn.phonetik.uni-muenchen.de/diagnostik_therapie/index.html)

The University of Sydney

Page 44

## Which assessments to prioritise? (Longley et al., 2012)

Neuropsychology assessment priority list.

1. **HIGH RISK OF DANGER TO CLIENT'S (OR CLIENT'S FAMILY'S) HEALTH OR SAFETY:** There is an urgent need to establish the role of possible cognitive or behavioural problems in any perceived health risks, so that appropriate safety measures can be implemented immediately.
2. **UNEXPECTED REHABILITATION (OR RESPITE STAY) MANAGEMENT DIFFICULTIES:** There is an urgent need to establish the role of possible cognitive or behavioural problems in any unexpected difficulties with the client's on-site rehabilitation (or 'Respite Stay') management, so that appropriate changes can be implemented immediately.
3. **URGENT LEGAL ISSUES REQUIRING NEUROPSYCHOLOGY INPUT:** There are significant legal processes (Guardianship application, property settlement, etc.) pending, which require formal documentation regarding the cognitive status of the client before they can proceed.
4. **IMPENDING FAMILY / SUPPORT/ ACCOMODATION / SERVICES RELATIONSHIP BREAKDOWN:** There is a fairly urgent need to gain an understanding of the role of possible cognitive or behavioural problems in the client's family or relationship difficulties, so that the client and family can be given appropriate management advice as soon as possible.
5. **IMPENDING LOSS OF CLIENT'S JOB:** There is a fairly urgent need to gain an understanding of the role of possible cognitive or behavioural problems in the client's ability to perform their job so that the client or employer can implement appropriate compensatory management strategies.
6. **CLIENT WANTING TO FIND WORK, OR TO CHANGE CURRENT WORK CONDITIONS:** There is a fairly urgent need to clarify the role of cognitive or behavioural changes in the client's ability to perform a specific job, so that appropriate work conditions can be organised.
7. **MAJOR CHANGES PREDICTED IN THE CLIENT'S LIFESTYLE / REHAB PROGRAMME:** There is a moderately urgent need for detailed information regarding the current cognitive or behavioural status of the client before major changes are / can be made to their lifestyle (such as in their rehabilitation program, living arrangements, enrolling in further study, etc.).
8. **CLIENT TRAVELLING IN FROM A DISTANCE FOR A REHABILITATION PROGRAMME:** There is a need for neuropsychological input into the rehabilitation programme of a client who is planning to travel in from a distance (probably NSW Country or ACT) for a limited stay.
9. **CLIENT (OR CLIENT'S FAMILY) EXPRESSING GENERAL CONCERN ABOUT COGNITIVE OR BEHAVIOURAL CHANGES:** There is a need for the client (or their family) to understand the nature and extent of any cognitive or behavioural changes so that they can manage their lives better in general.

Page 45

## Assessment of everyday communication and impact on function

## How to assess impact of PPA on everyday function

- **Interview** pwPPA and family member:
  - **What's causing you most difficulty in your day to day life?**
- **Observe** language use, communication strategies in clinic
- **10-minute conversation sample with communication partner:** reveals trouble-indicating behaviours, repair strategies and success
- **Oromotor assessment with cranial nerve assessment:** impairments predictive of nvPPA and swallowing difficulties
- **Fluid trial:** Offer pwPPA a cup of tea, observe and ask about swallowing, provide education about how to monitor changes in swallowing and minimise risk
- **Scales and questionnaires:** impact on functional communication, coping, mood (PwPPA and partner)
- **Nutrition screen**

The University of Sydney

Page 47

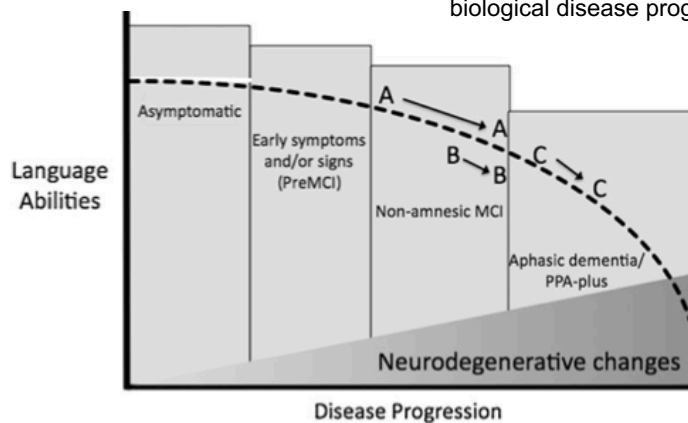
## Review Assessment: Assessing change over time

The University of Sydney

Page 48

## Rate of decline varies between individuals

Sapolsky et al., 2011, Figure 1  
The clinical trajectory of primary progressive aphasia as a function of biological disease progression

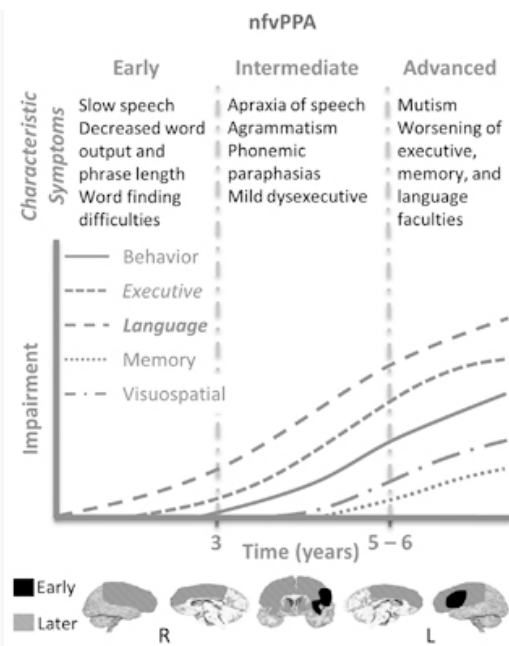


The University of Sydney

Page 49

## Clinical hypotheses about change over time: nvfPPA

Karageorgiou & Miller (2014, Fig. 2)

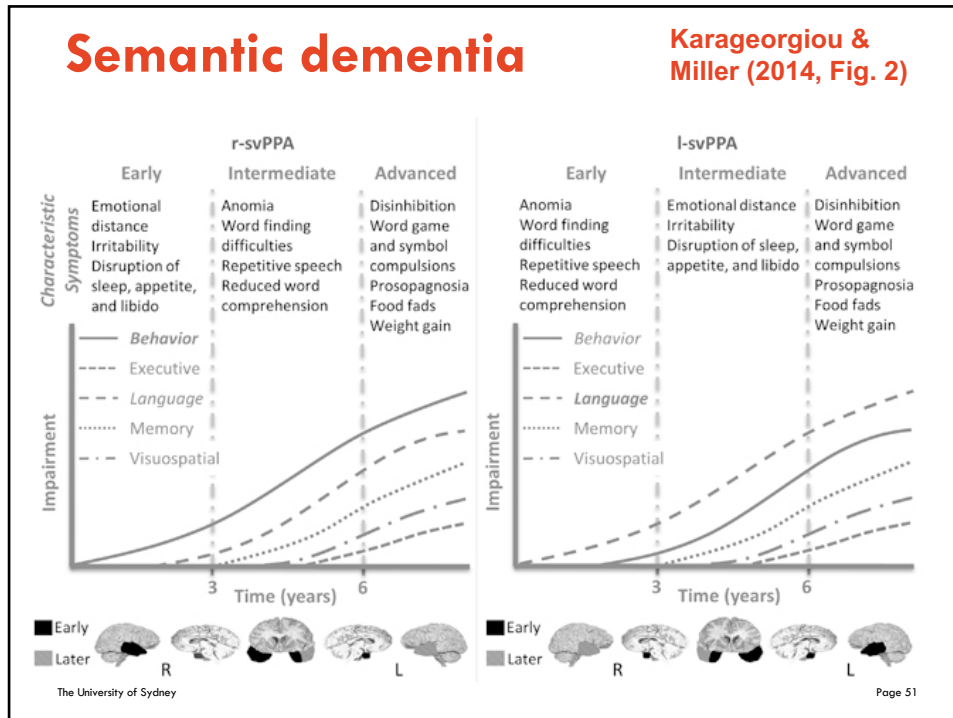


The University of Sydney

50

## Semantic dementia

Karageorgiou &  
Miller (2014, Fig. 2)



## Progressive Aphasia Severity Scale PASS (Dickerson, 2011)

### Rating scale designed to measure change over time

Rates performance and severity of impairment based on standardised tests, questionnaires, and functional communication (0=normal to 3=severe impairment) in ten language domains:

1. Articulation
2. Fluency
3. Syntax and grammar
4. Word retrieval and expression
5. Repetition
6. Auditory comprehension
7. Single word comprehension
8. Reading
9. Writing
10. Functional Communication

More information about PASS, including questionnaires and training materials can be found at <http://www.ftd-boston.org/>

## Rapid decline: TOC

### Summary of Case History

**June 2012** Initial assessment and diagnosis of nvPPA based on effortful speech production with inconsistent articulation errors, disturbed prosody, word finding difficulties agrammatism, and orobuccal apraxia

**November 2012** Modified Barium Swallow- adequate oropharyngeal swallow with mild residue in pyriform fossa and valleculae. Continues on normal diet with strategies to minimise risk. European holiday: asked neurologist to provide letter in case of need for medical care whilst overseas. Education for partner. Referred to dietitian.

**April 2013** Stops Aricept because of swallowing difficulty. Other medications crushed with yoghurt/custard. Still physically active, playing tennis, yoga, swimming: good for chest status and alertness.

**May 2013** USA holiday

**June 2013** Hospital admission for bladder repair. Speech language review one week later: mute, significant weight loss and throat clearing for trace aspiration

## Rapid change over time: TOC

### Transfer to residential aged care

- **July 2013** -Review: mute, at times confused and then distressed, presence of stereotypical behaviours, worsening dysphagia, soft diet, dysphagia cup, repeat MBS but still no frank aspiration observed. Recommended modified minced diet and thin fluids with safe swallow approach. Further education to TOC re choking hazard, safe strategies.
- **November 2013**- residential aged care, education of care staff, handover to nursing staff.
- **December 2013** - handover to private Speech Pathologist: ongoing monitoring of hydration, balancing thickened fluids vs distressing coughing in accordance with TOC's preferences during palliative care, managing secretions
- TOC appreciative of care

## Rapid change over time: TOC

### What we learned:

- Increased vigilance for rapid deterioration
- Monitor weight
- TOC quickly became adynamic, didn't initiate communication so AAC was less helpful
- She was able to take 2 much-loved overseas trips. Letter from doctor explained her condition and needs in case of deterioration
- Partner and wide network of friends collaborative in TOC's care: Education of family and management of dysphagia (swallowing difficulty) allowed palliative care to commence at home, extending the time TOC was able to remain at home

## Planning for discharge from Speech Pathology services



## Planning for discharge

- pwPPA report feeling abandoned when discharged and see it as a sign of no further hope
- Described by Hersh (2009) in acute onset aphasia
- Discharge may not be appropriate given changing needs with progression: Recommend ongoing review with ongoing support and education
- Constrained by service provider's guidelines
- Australian context: form relationship with primary health care provider (e.g. GP) and maintain that relationship and communication throughout the patient journey
- **Who has ongoing responsibility for case management?**

The University of Sydney

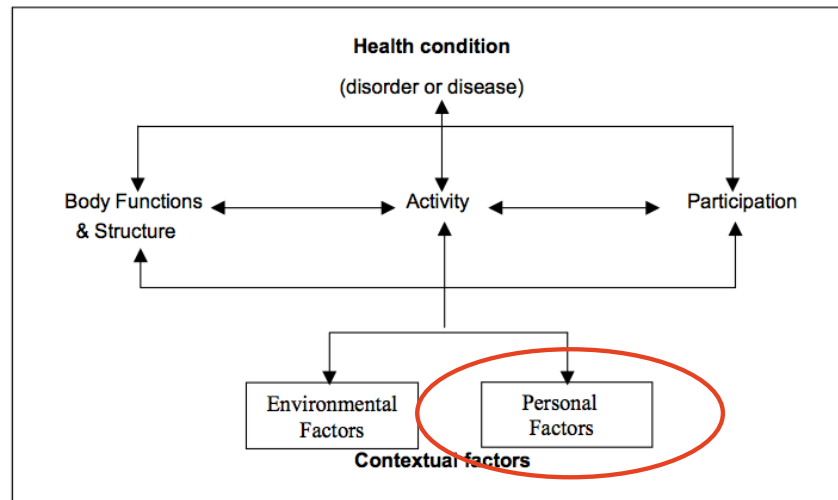
Page 57

## Assessing contextual factors

The University of Sydney

Page 58

## Assessing personal factors: pw PPA



The University of Sydney

Page 59

## Assessing personal factors

### Observe and ask about emotional wellbeing

- Anxiety, depression including suicidality
- Ask an informant

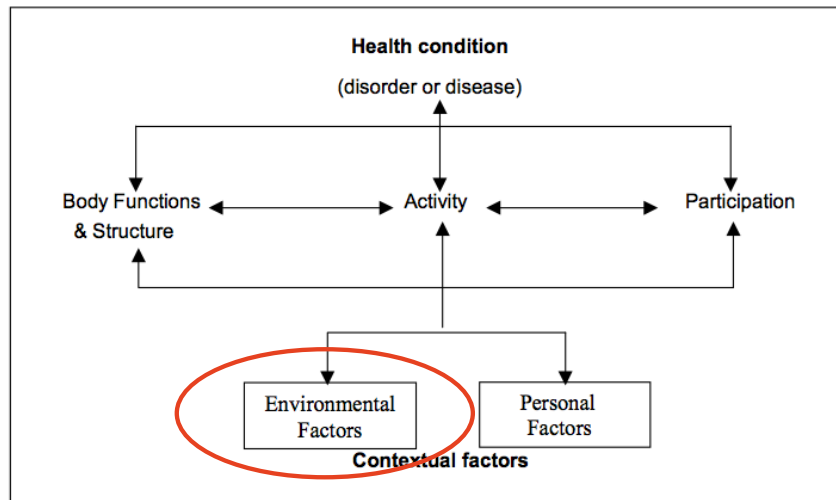
### Coping strategies

- Brief Cope: Withdrawal and emotion-focused coping items predicts poorer well-being in carers (Roche et al., 2015)

The University of Sydney

Page 60

## Assessing environmental barriers and supports



The University of Sydney

Page 61

## Assessing environmental factors

### Partner, family members, significant others

- Observe and ask about physical health
- Observe and ask about emotional well-being and coping
- Carers: Ask about positive experiences of caring as well as difficulties and burden.
- Be alert to needs of young adult children in younger onset cases

### Access to care

- Mobility limitations/Transport difficulties of pwPPA
- Cultural/language barriers amplified by language impairment
- Resources of healthcare provider: number of sessions, experience of clinician

The University of Sydney

Page 62

## The healthcare team

### Consider further referral to

- Speech Pathologist
- Clinical psychologist/Family therapist
- Social worker
- Occupational therapist
- Physiotherapist/Exercise physiologist
- Dietician

### Note unattended/unsolved medical problems and refer

- Major cancer
- Urinary problems
- Cough, sore throat

## Intervention & management

Feedback and goal-setting

Disease-modifying interventions

Interventions targeting speech and language impairment

Interventions targeting communication activities and life participation

Interventions targeting personal and environmental contextual factors

## **Feedback and goal setting**

## **Feedback as the first intervention**

- The “bridge to intervention”
- Can be therapeutic in itself
- Indicates weaknesses and strengths
- Contributes to collaborative goal-setting, choice of interventions, planning for the future
- Influences motivation
- Provide written information to support comprehension and memory in aphasia-friendly format

## Feedback as the first intervention

### Neuropsychological assessment with feedback as an intervention for people with Multiple Sclerosis (Longley et al., 2012)

- Set within therapeutic neuropsychological assessment framework
- Refundable in Australia as psychoeducational intervention
- Ask at the beginning: "How much do you want to know about the results?"
- Clinician needs to be able to sit with the discomfort of breaking bad news
- Takes time (1+ hour)
- Involve person with MS and family

The University of Sydney

Page 67

Yes/no	Issue/Topic	Notes (e.g., why topic not covered)
	<b>Feedback conditions:</b>	<b>Longley et al., (2012)</b> <b>Checklist for feedback session</b>
<input type="checkbox"/>	Client was accompanied by a caregiver or other(s)	
<input type="checkbox"/>	Attendees agreed for session to be audio-recorded	
	<b>Essential feedback components:</b>	
<input type="checkbox"/>	Note if Neuropsychologist had advised client to bring a caregiver	
<input type="checkbox"/>	Reviewed general purpose of assessment	
<input type="checkbox"/>	Reviewed client's original views/concerns	
<input type="checkbox"/>	Reviewed caregiver's (or other's) original views/concerns	
<input type="checkbox"/>	Explained the assessment process (including the tests, use of norms, basic test interpretation)	
<input type="checkbox"/>	Explained test results and observed behaviours	
<input type="checkbox"/>	Described client's cognitive strengths and weaknesses	
<input type="checkbox"/>	Discussed likely causes of cognitive profile	
<input type="checkbox"/>	Addressed issues of low self-esteem (related to Neuropsychological assessment process or assessment results)	
<input type="checkbox"/>	Explained likely real-life consequences of cog imp (including addressing the referral question)	
<input type="checkbox"/>	Provided general, strategic, cognitive impairment management advice	
<input type="checkbox"/>	Provided specific management recommendations for client to f/u	
	<b>Optional feedback components, if appropriate:</b>	
<input type="checkbox"/>	Discussed prognosis (i.e. likely cognitive change over time)	
<input type="checkbox"/>	Provided specific management recommendations for caregiver to f/u	
<input type="checkbox"/>	Provided advice on other issues [*List issues over page]	
<input type="checkbox"/>	Recommended referring on to other health pros	
<input type="checkbox"/>	Provided notes for client &/or carer to take away	
<input type="checkbox"/>	Audio recorded the feedback session & promised to burn onto CD and send to client &/or carer ASAP	
<input type="checkbox"/>	Offered a f/u feedback and discussion session	

Page 68

## **Treatment planning and goal setting (Volkmer, 2013, Chapter 3)**

- Identify need, choose intervention to respond to need
- Understanding that a slowing of deterioration, stability or improvement may all be positive outcomes in PPA
- Specific Measurable Attainable Relevant Timely (SMART) goals that describe outcomes you want to achieve
- Goal needs to be specific, relevant to pwPPA and family, and achievable to motivate initial engagement and treatment adherence
- Measuring outcomes: Goal Attainment Scaling
- Impairment-directed treatment typically results in positive change
- Suggested questions for goal-setting, examples of goals, using goals to form therapy plans: see Volkmer, (2013)

The University of Sydney

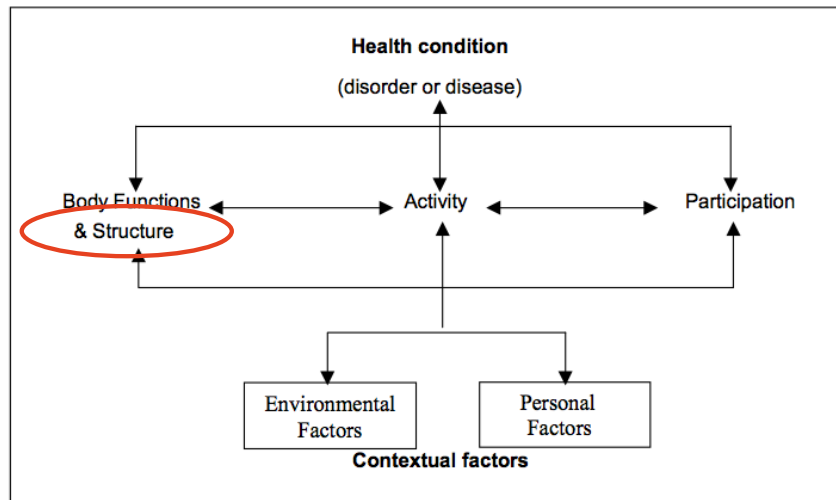
Page 69

## **Disease-modifying interventions**

The University of Sydney

Page 70

## Disease-modifying interventions



The University of Sydney

Page 71

There are currently no treatments to stop or slow the degenerative process and there are only very limited medication options for the management of the cognitive-behavioural symptoms. However, alternative, non-pharmacological interventions may offer significant benefit to the quality of life of the diagnosed individual.

(Kortte & Rogalski, 2013, p. 237)

The University of Sydney

Page 72



## Pharmacological intervention

(Karageorgiou & Miller, 2014)

Table 5 Pharmacological treatments<sup>64-82</sup>

Medication	Dose	Population	Study designs	Combined study outcome	Side effects
Trazodone	Up to 300 mg daily	bvFTD	DB-CO-RCT	Improved behavior <sup>a</sup>	Fatigue, dizziness, hypotension
Fluvoxamine	50–150 mg daily	bvFTD, svPPA	OL	Improved stereotypies	Appetite loss
Paroxetine	Up to 40 mg daily	bvFTD	OL, OL-RCT, DB-CO-RCT	No definite behavioral benefit Improved mood, compulsions, and eating disorders	Well tolerated
Fluoxetine	20 mg daily	bvFTD	OL	Improved mood, compulsions, and eating disorders	Well tolerated
Sertraline	50–125 mg daily	bvFTD	OL-CT, OL	Improved stereotypies	Well tolerated
Citalopram	40 mg daily	bvFTD	OL	Improved behavior	Well tolerated
Donepezil	Up to 10 mg daily	bvFTD	OL, DC	No benefit	Worse behavioral symptoms
Galantamine	Up to 24 mg daily	bvFTD, PPA	OL to DB-RCT	No benefit	Mild GI symptoms
Rivastigmine	Up to 9 mg daily	bvFTD	OL-CT	Improved behavior	Well tolerated
Quetiapine	Up to 150 mg total daily dose	bvFTD, nvPPA, svPPA	DB-CO-RCT	No definite benefit	Somnolence
Olanzapine	Up to 10 mg daily	bvFTD	OL	Improved agitation and anxiety	Somnolence, mild GI symptoms
Bromocriptine	Up to 7.5 mg 3 times daily	PPA	DB-CO-RCT	No benefit	Rare frustration intolerance
Methylphenidate	40 mg once	bvFTD	DB-CO-CT	Improved decision making within a few hours	Non-significant blood pressure increase
Dextroamphetamine	20 mg total daily dose	bvFTD, nvPPA, svPPA	DB-CO-RCT	Improved behavior	Well tolerated
Mefenamine	Up to 20 mg daily	bvFTD, nvPPA, svPPA	OL, DB-RC	No benefit	Well tolerated

Abbreviations: CO, crossover; CT, control trial; DB, double blind; DC, discontinuation of treatment; GI, gastrointestinal; OL, open-label; RCT, randomized control trial.  
<sup>a</sup>Improved behavior usually corresponds to neuropsychiatric inventory scores or refers to irritability, agitation, depression, and eating disorders.

The University of Sydney

Page 73

## Brain health for pwPPA and partner

**Address lifestyle factors to improve cerebrovascular function and reduce risk/effects of co-morbid disease, improve quality of life**

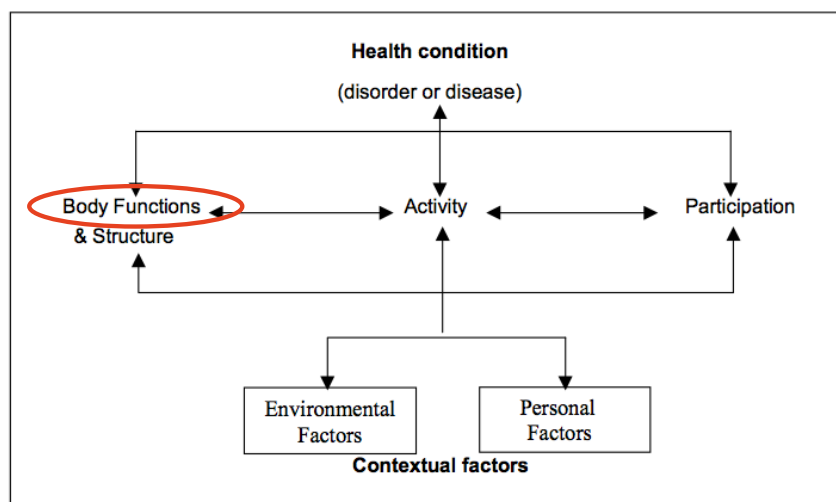
- **Quit smoking**
- **Manage obesity, high blood pressure, high cholesterol, diabetes**
- **Alcohol** – within limits advised by medical professional
- **Exercise**
  - Under medical advice
  - Mix of aerobic, flexibility, strength and weight-bearing exercise
  - Step counter
  - 30 mins per day
- **Relaxation**
  - Breathing, progressive relaxation and mindfulness
  - Stimulating/pleasant activities **“I plan a visit for Mike and I, to an exhibition or gallery, each week. Its good for both of us!”** (Lynne, wife of pwPPA)

## Interventions targeting speech and language impairment

The University of Sydney

Page 75

## Interventions targeting speech and language impairment



The University of Sydney

Page 76

## Review of evidence base for speech-language intervention in PPA

- Several reviews in the recent years, including a systematic review of nonpharmacological interventions for PPA (Carthery-Goulart et.al., 2013)

**Treatment effects:** consistently demonstrated

- Significant improvement in naming trained items for all participants across all studies

**Maintenance:** Gains lost within 2-6 months of ceasing therapy activities  
– person needs to continue therapy activities to maintain gains

**Generalisation**

- 17 studies include measures of generalisation to untreated items
- 6 showed within-level generalisation
- Across-level generalisation more difficult to demonstrate due to sample size, discourse genres, variation in measures.
- Recent results targeting connected speech are promising (Beales et al., 2016)

## Choice of treatment when there is a limited evidence base

- Use clinical judgement to apply **treatments used in other areas of SLP practice** (aphasia, apraxia, motor speech disorder e.g. LSVT, partner training, dysphagia, AAC, education & feedback) where there may be evidence base from other populations (McNeil & Duffy, 2001)
- Similarly, **practices effective in dementia care** may be adopted especially in more advanced disease or where behavioural symptoms are present, e.g. behavioural management of BPSD (clinical psychologists, OT, geriatrician), carer support & education programmes, memory/life story books)
- **Expert opinion papers** e.g. Kindell et al. (2015) semantic dementia, O'Connor et al., (2014) role of occupational therapist

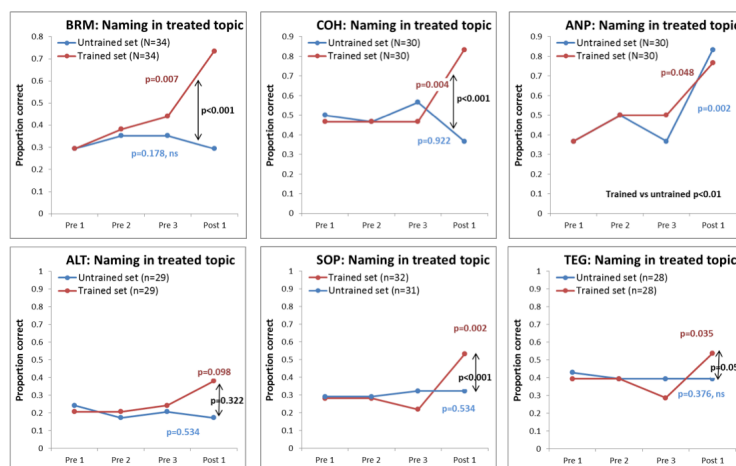
## Word retrieval treatment



The University of Sydney

Page 79

## Lexical retrieval treatment: Reliable treatment effects



Croot, Nickels, Taylor et al. (in prep.)

Page 80

## Word retrieval treatment

**Jokel et al.,  
2011**

- > 3 Ps with fluent PPA
- > 10 hours training
- > 80 words
  - 20 semantic
  - 20 phonological
  - 40 control

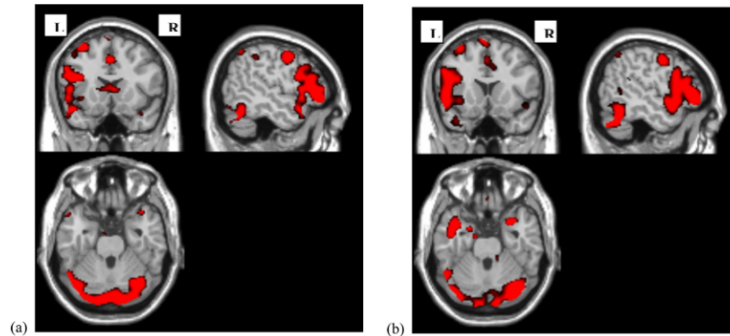


Figure 1. Activation in anterior temporal lobe (a) pre-treatment, (b) post-treatment in CW

**Pre-Treatment**

**Post-treatment**

The University of Sydney

*Alzheimer's & Dementia*, 2011, p. S754

Page 81

## Word retrieval treatment

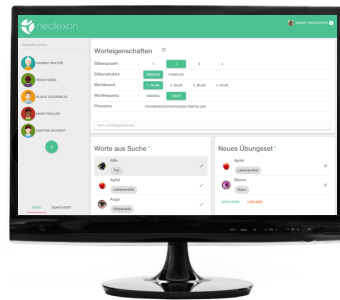
Evidence-based list of factors that may facilitate therapy (Jokel et al., 2014)

- Greater success is achieved with personally familiar items (Green Heredia et al., 2009; Jokel, Rochon, et al., 2006; Snowden & Neary, 2002; Snowden et al., 1994, 1999).
- Organising items into semantic categories may improve relearning and retention (Graham et al., 1999).
- Involvement of the client in item selection facilitates motivation (Jokel, Rochon, et al., 2006).
- Better success is possible with at least partially spared semantics in svPPA (Jokel, Rochon, et al., 2006; Snowden & Neary, 2002).
- Home-based practice is an effective way of improving vocabulary (Bier et al., 2009; Green Heredia et al., 2009; Savage et al., 2013).
- Practice is helpful to maintain existing vocabulary, regardless of the PPA variant (e.g., Green Heredia et al., 2009; Jokel & Anderson, 2008).
- Rehearsal of known items delays their decline (Jokel, Rochon, et al., 2006; Newhart et al., 2009).
- Incorporation of re-trained words into daily communication aids their maintenance and generalisation (Green Heredia et al., 2009; Jokel, Rochon, et al., 2006, 2010).
- The errorless approach is more successful than the traditional errorful approach in svPPA (Jokel & Anderson, 2012; Jokel et al., 2007).
- There is some evidence that phonologically based treatment may be more beneficial for nvPPA, while semantically based therapy may be equally or more successful for svPPA (Louis et al., 2001); but there is also evidence that both phonological and semantically based therapy may be successful in the same patient with PPA (Jokel & Anderson, 2010, 2012).
- Computer-based treatment for anomia is appropriate for mild-to-moderate PPA (Jokel et al., 2009).
- Magnetic brain stimulation has shown some promising results, but further studies are needed (Finocchiaro et al., 2006; Trebbastoni et al., 2013).
- Better success is possible with at least partially spared semantics in svPPA (Jokel, Rochon, et al., 2006; Snowden & Neary, 2002).
- Improvements resulting from behavioural treatment may induce increased activation (measured via neuroimaging methods) of both affected and spared brain regions (Dressel et al., 2010; Jokel et al., 2011).

The University of Sydney

## A therapy software for individual word training

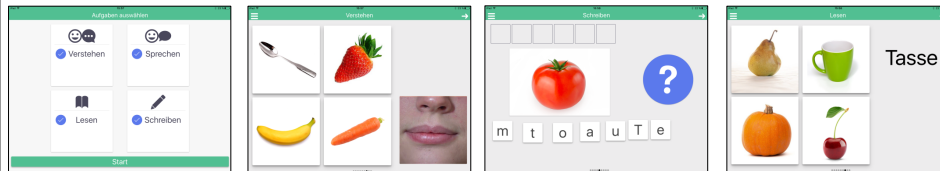
www.neolexon.de (newsletter subscription)  
info@neolexon.de



Therapist configures an individual set of training words out of 10.000 words

Tailored set of words can be practiced on the tablet in therapy sessions (Therapy App) and at home (Home App)

Therapist tracks the performance from the results visualizations



## Treatment targeting communication activities of daily living

### Script training

- Aims to improve communication in specific situations that are important and challenging for the pwPPA
- pwPPA rehearses and learns personally-relevant phrases

### Scaffolding

- Also aims to improve communication in situations selected by pwPPA
- Identify lexical items required and provide practical supports (e.g. cards, notebook, practice on the way there, word relearning)

**Rich personal picture descriptions** (Kindell et al., 2015)

## Treatment for speech production and fluency in nonfluent/agrammatic PPA (Henry et al., 2016)

- Video-Implemented Script Training for Aphasia (VISTA)
  - Adapted from **speech entrainment** technique (Fridriksson et al., 2012) and VAST application (<http://www.speakinmotion.com>)
  - At-home script training practice (30 minutes per day) with audio-visual model
  - Sessions with clinician twice per week (45 min. each) to promote memorization and conversational usage
- N=10 have completed treatment to-date
  - N = 5 in-person treatment
  - N = 5 teletherapy
- Follow-up testing at 3 mo, 6 mo, 1 year post-treatment

The University of Sydney

Page 85

## VISTA (Henry et al., 2016)

### Participants

- Individuals with nonfluent/agrammatic PPA in the mild to moderate stages
- Linguistic or motor speech impairment greater than other cognitive impairments
- Intact repetition of at least five syllables
- MMSE score of 15 or greater

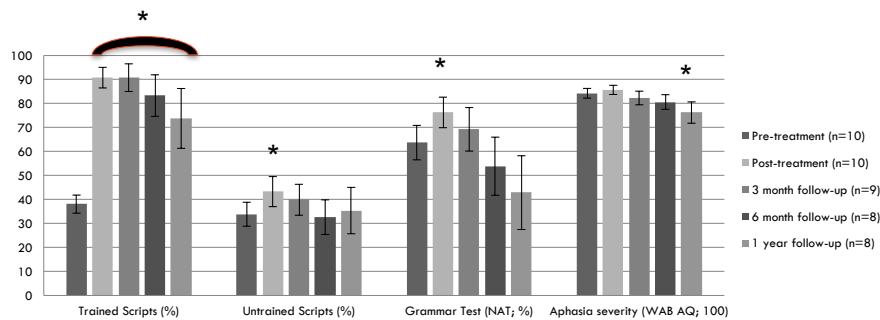
### Script development

- Collaborative with patient, family and clinician
- Scripts are
  - Personally relevant
  - Tailored to the individual's motoric and linguistic profile
    - Speech rate for stimuli derived from Ps reading rate

The University of Sydney

Page 86

## VISTA outcomes to-date (Henry et al., 2016)



\*=  $p < 0.05$ , Wilcoxon, relative to pre-treatment score

Primary outcome measure = % correct, intelligible words

The University of Sydney

Page 87

## Constraint Induced Aphasia Therapy for verb phrase and sentence structure (Hameister et al., 2015)

- **CIAT** (Pulvermüller et al., 2001)
  - well-studied and effective treatment in stroke-induced aphasia
  - based on the theory of „learned nonuse“ (Taub et al., 1989)
  - successful modification with regard to different types and severities of aphasia (Meinzer et al., 2012)
- **3 main principles**
  - Massed Practice
  - Constraints & Shaping
  - Reinforcement

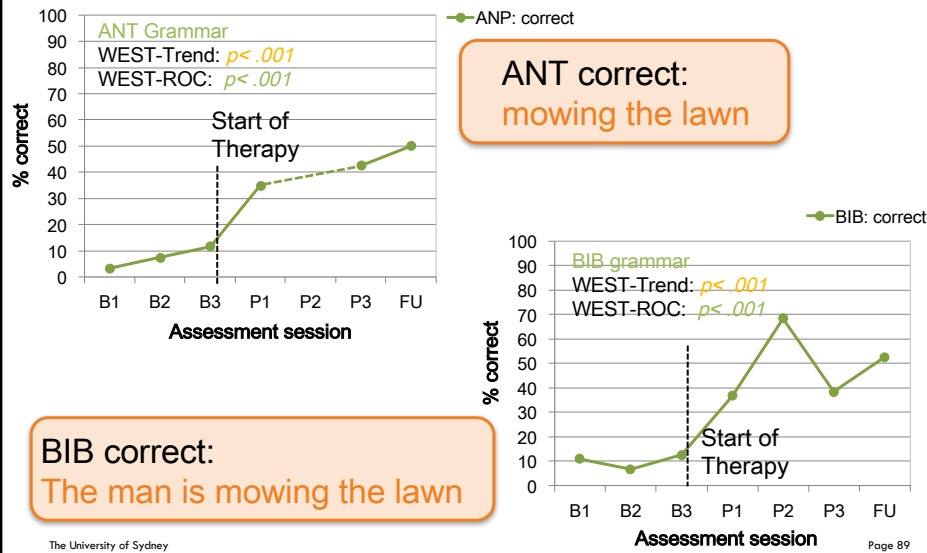


The University of Sydney

Page 88



## Grammatical completeness



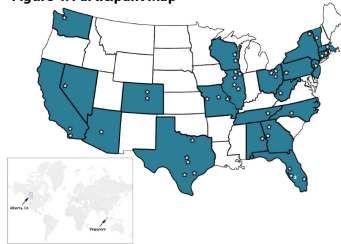
## Brain stimulation

- to augment treatment effects
- inhibit or excite neuronal function, depending on whether the stimulation is low versus high frequency
- rTMS (e.g. Finocchiaro et al., 2006)
- tDCS (e.g. Tsapkini et al., 2014)
- Approaches are currently experimental with promising results

## Telemedicine (Rogalski et al., 2016)

# communication BRIDGE

Figure 4. Participant Map



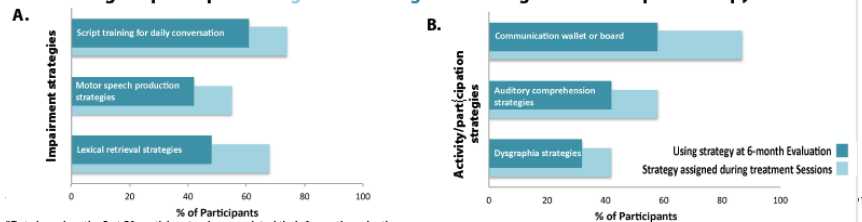
### Impairment strategies

- Script training
- Motor speech training
- Lexical retrieval

### Activity/participation strategies

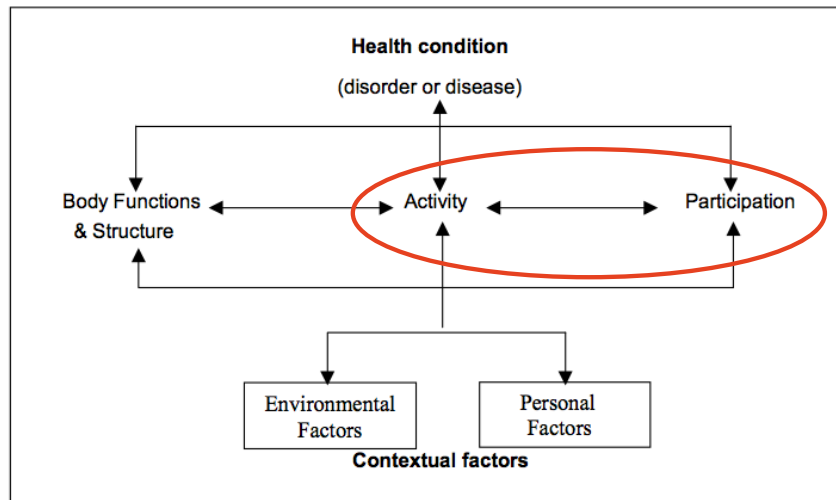
- Personalised communication wallets/boards
- Auditory comprehension strategies in conversation
- Dysgraphia strategies

### 5. Percentage of participants assigned and using their strategies 6-months post therapy\*



## Interventions targeting communication activities and life participation

## Interventions targeting communication activities and life participation



The University of Sydney

Page 93

## Interventions targeting communication activities and life participation

- Communication partner training
- Groups
- AAC
  - Writing and reading
  - Speaking devices
  - Communication boards/mats
  - Communication, life-story and memory books

The University of Sydney

Page 94

**“We don’t communicate in single words”**

**....Conversation**



The University of Sydney



## **Conversational trouble in PPA**

**(Taylor et al., in prep; 2014)**

- Trouble-indicating behaviours were interactive (asks for help from partner) and non-interactive (non-continuation, fillers)
- Two people with nvPPA had sparse output but equal turn-taking with interactive trouble-indicating behaviours
- Seven people with svPPA were also active participants in conversation, but experienced a great deal of trouble in conversation. Wide variation among individual couples.
- Communication behaviours of the conversation partner were critical to conversational success

The University of Sydney

Page 96

## Conversation training

- Review conversation sample for facilitative and non-facilitative behaviours
  - Couple training or group training
  - Empower pwPPA and partner to negotiate strategies
  - Training slides to educate partner
  - Written handouts
  - Avoid judgmental tone
  - Encourage partner reflection and specific goal setting
  - Video to train partner?
- Experience of colleagues training communication partners of pwTBI:
- Better to use anonymous examples
  - Role play with colleague

### Facilitative

- Prepare the environment
- Use alerters
- Slow rate
- Reduce syntactic complexity
- Reduce message length
- Increasing redundancy
- Cognitive support-gives information when needed; uses memory, organization supports; gives cues in a conversational manner; responds to errors by giving correct information in a non-punitive manner
- Emotional support-  
**“We’re in this together!”**

### Non-facilitative

- Long word search
- Testing questions
- Repetition without additional information
- Punitive correction

## Avoiding testing questions

E.g. Tell the doctor  
what we did  
yesterday



**“ I find the best way to stop students from talking to each other is to keep asking questions. Then they have nothing to say.”**

The University of Sydney

## Avoiding testing questions

Tell the  
doctor what  
we did  
yesterday

**Testing question**

Didn't we have  
a lovely day  
yesterday?  
Down at the  
harbour?

**Reveals competence**

The University of Sydney

Page 100

## Encouraging partner to increase facilitative behaviours or decrease non-facilitative behaviours



### Examples of goals

1. Increase pause time
2. Provide more context
3. Eliminate testing questions
4. Encourage gesture
5. Encourage use of written words or part words



### Multiplier effect

- Family members, close friends will mirror how you interact with your partner

## Empowering pwPPA and partner to negotiate strategies

**Wife of man with PPA:** “The most helpful thing was your suggestion to negotiate between us how to deal with it when Keith can’t think of the word.”

**Man with PPA:** “Yeah – I don’t feel like the little boy in the corner any more.”

## Conversational strengths: Introducing WIC

- 75 year old male
- formerly an airline pilot with 13+ years of education
- right predominant svPPA
- ACE-R 72/100



## Difficulty discussing prognosis

- Partner avoiding talk of prognosis:  
6/7 partners (n=10) did not engage when the topic was prognosis/what will happen in the future (Taylor et al., 2015 and in prep.)
- NB may be because on video

## Harmonizing Together

Speech and Music Therapy and Support for Patients and Partners With Primary Progressive Aphasia

Katie Brandt, MM  
Community Resource Specialist

Megan Quimby, SLP  
Speech and Language Pathologist

Frontotemporal Disorders Unit  
Massachusetts General Hospital  
Boston, Massachusetts, USA  
[www.FTD-Boston.org](http://www.FTD-Boston.org)



## Alternative augmentative communication (AAC)

**Acceptability:** People may want to continue working on spoken language

- E-technologies in phone/i-pad/tablet apps more acceptable than older electronic technologies
  - E.g. Phone: take photographs to scaffold storytelling
  - Many new apps available: guide clients in purchase
- Writing on notepads, in notebooks, diaries etc. often adopted spontaneously
- For communication partners: write as you speak

The University of Sydney

Page 107

## AAC: Notebooks, diaries, writing

- Often adopted spontaneously by person with PPA
- Used to support comprehension, production, life participation

### October Year 11

41. Woche 284 - 81	October Octobre	Thursday Jeudi	11.
	October	Donnerstag	
It's a beautiful day, then Bonne vie heute. Wunderbar!	+ 13°C / in der Sonne + 15/16°C / mittags		7
Um 9:30 Uhr stehe ich auf + esse dann "2" Morge...			
Am Nachmittag muss ich um 15:30 Uhr in die Orthopädie von ... gehen.			
Ich gehe dann alleine in S. Post.			
Da ich 1 Tag frische, ich bleibe um 15 Minuten.			
Am Abend dann habe ich um 16:4 Uhr			
etw. + dann fahre ich in die Mitternacht			
um 17:30, dann fahre ich ab + fahre um 17:45 Uhr wieder ein.			
Das ist ein Tag, wo wir Einkäufe machen von ...			
Das ist ein Tag, wo wir Einkäufe machen von ...			

The University of Sydney

### December Year 12

29. Monday Lundi	December Décembre	1. Woche 36
Montag	Dezember	
Schönes Wetter, Reise W/etken	- 5°C / in der Sonne - 10°C / mittags	
Um 9 Uhr stehe ich auf + dusche + trinke mich in dem Wuschbecken. Ich räume mich + putze mich. Amontel/Wochen im Badzimmer, nachdem ich um 2 "2" Morge" + ich sage bei mir viele Zeitungen. Ich um 11 Uhr fahre ich zum Zahnarzt mit einem ...		
Am Nachmittag - das ich viel + erhalte von ...		
Am frühen Nachmittag. Gegen den Abend ...		
Ich ... in der Küche + auf dem Rücken - hand ist in der Küche. Auch um ... stellen fest, dass die Tage - auch in der Schweiz - immer mehr länger werden.		

14

Heitkamp et al., 2016

## Boyd and Shirley: The Lightwriter

### THE LIGHTWRITER

I had been keeping in touch with a speech pathologist in Sydney by email and telephone. She was the only person I had found that knew about PPA besides the neurologist. I had been finding it hard to know what to do for Boyd, how I could help him to make things easier for him. She suggested that a device called a Lightwriter could be helpful for some people. I was very interested in it. I spoke to Boyd about it and he was keen to try it if we could get one. He was still working on the computer now and again: He was able to type sentences and understood how the computer worked. He liked downloading our holiday photos from the camera and saving them on discs.

Boyd was now visiting a speech pathologist in a city one-and-a-half hours away, who was helping him with speech exercises. She thought the Lightwriter would be good for him. She made enquiries to find one, and then made an application to a government department giving details of her assessment that Boyd's speech was very poor, and that an alternative communication device, specifically a Lightwriter or similar, would enable him to communicate more. The government department said they could not supply this appliance. It was a big disappointment. I wanted him to have the chance to use one because I felt he would be able to work it.

I got in touch with the Welfare Officer at the local Returned and Services League (RSL) club. He applied to the government department again, but he was unsuccessful as well. As he was the president of the club, he called a meeting and they decided to buy the Lightwriter for Boyd. It was a great thing for them to do and we were so pleased. Boyd had it within a week, in June 2009.

Boyd worked it out quite easily and was pleased with what he could do with it. The Lightwriter had a memory, so he put in all the different things he would ask regularly and greetings for different people that he knew by name. He would press a certain letter or number that he had keyed in for each sentence and it would come up. If someone asked him something, he was able to type in the answer. The Lightwriter had a two-sided display, so he could see what he was typing in and the person he was talking to could also see it. It gave Boyd his quality of life back again and he had confidence now to talk to people through the machine. It was definitely the right time for him to have it while he still had the cognitive skills to use it.

He settled down again from being frustrated and agitated and was happier in himself for a while. We went away on some day trips and short stays to catch up with family and friends. Boyd said he liked to travel about.

The University of Sydney

### MORE CHANGES

Boyd's speech was starting to get less and less. He was just getting odd words out now. The local speech pathologist couldn't help much more, but we went to see her a few more times. She wanted to know if Boyd was having any problems swallowing, which at this stage he wasn't. She was happy that he was using the Lightwriter well. She made up some sheets for him with pictures and sentences on them for asking for things that he wanted and would like to do. I put them in a book for when he would

OUR JOURNEY WITH PPA 903

need to use them, and I put in pictures of food he liked to eat—desserts and fruit especially! I also wrote down the little jobs he liked to do around the house.

I was realising that Boyd was trying to keep up all the things that he was used to doing as long as he could. He wasn't in his workshop as much now: He was starting to have difficulty using his tools to make things. He liked pottering in the garden though. He still wanted to drive, but his movements had become more unstable in the car and he wouldn't let me drive. When we saw the neurologist again, I told him about Boyd's movements in the car. I was very upset to have to do this, but I was getting nervous about the way he was driving. We had a lot of disagreements about it. The doctor told Boyd he should not drive anymore. Boyd took notice of the doctor straight away and told me to do the driving from then on. He was withdrawn for a few days, but never tried to drive again. He was just a back-seat driver for a while!

We always discussed everything together. He could still communicate quite well on his Lightwriter at this time. I would never do anything to do with our affairs without talking to Boyd first. I also would ask him what he would like to do or places he would like to go. I did my best to make sure that was always the case, even later on when his skills worsened.

Boyd was managing with the Lightwriter well, which was good because his speech was getting worse. He was always doing things, keeping himself busy and seemed to have accepted the fact that he had this rare disease. He still had the jerky movements at times. He went along quite well for a few months.

Page 109

## Voice, message and story banking

**Voice banking** - special software used to record a large sample of person's speech that is consolidated to create a synthetic voice approximating their natural speaking voice.

**Message banking** - using a digital recorder/computer recording software to record and store words, phrases, sentences, and personally meaningful expressions in the person's natural speaking voice while it is still intact

**Story banking** - similar to message banking focussed on recording favorite stories. These are personal, legacy stories that are available for sharing with others later, when speech has become more difficult

The University of Sydney

Page 110

## Boyd and Shirley: The communication book

### 2011: THE SIXTH YEAR

Boyd seemed to get very tired quickly and was sitting around in the chair more now. It didn't help that he was still up and down to the toilet frequently, which was becoming more of a problem, and the urologist prescribed medication for it. He agreed with the neurologist that it was related to Boyd's brain condition.

Boyd had stopped going down to his workshop. He wasn't doing anything in the garden except when he tried to mow the lawn a few times, but it had tired him out and I had to finish it. I talked to him about getting someone in to cut the grass, but he wasn't happy about it. I said it was getting too much for either of us to do, so I

OUR JOURNEY WITH PPA 905

arranged for the grass to be cut for us. My work load was getting heavier, so that was one less job to worry about.

Boyd's speech was very poor now, just an odd word here and there. He was very dependent on the Lightwriter now. He didn't try to type much, mainly used the memory. When he wrote something, it would be two or three words and you had to work out what he trying to say. This is when the book I had made up with pictures came into use every day. I left it handy for him to see. He then started using it himself.

He was trying to read books still, but I noticed he would be on the same page for quite a while. He always enjoyed reading the paper, but he wasn't reading it properly any more, and put it down after a little while. He was watching the TV a lot and liked watching western action movies. He could still put the television on himself and put DVDs on when he wanted to. He wasn't using the computer any more. He had very set routines that he could do and didn't forget how to do them, like going to the post box for the mail, setting the table, making morning tea and putting the bin out. He was having trouble remembering past events and peoples' names, except for the people he saw on a regular basis.

The University of Sydney

## Communication books

Rogers et al. (2000)

***“should be regarded as an essential communication tool for individuals with PPA”*** (p. 326)

BUT

***“clinicians frequently report that most recipients of prefabricated books rarely use them”*** (p. 326)

The University of Sydney

Page 112

## Successful Communication books (Rogers et al., 2000)

### Personalised

- Begin early, allowing the person with PPA to take lead role in selecting and organising content
- include pictures, symbols, photographs, written words, phrases, names

### Utilised

- Provide opportunities to use it
- Clinician observe conversations between person with PPA and partner for 10 minutes each week during therapy

### Dynamic

- It's a tool, not a photograph album or a "showpiece"
- The person with PPA should be adding to it without the clinician's input
- Partners adopt responsibility for adding to book with disease progression

## Life Books as a memory/ communication aid

- Systematic review by Egan (2010) investigated methods to improve verbal communication of people with AD
- The most effective way to improve communication was the combined approach of a **memory/communication aid** plus **caregiver training**.
- Enhance topic maintenance (time on topic, and words per topic with fewer topic changes). Assist in maintaining verbal attention, help individuals focus their thoughts.
- **My Life Story app** – recommended by colleagues

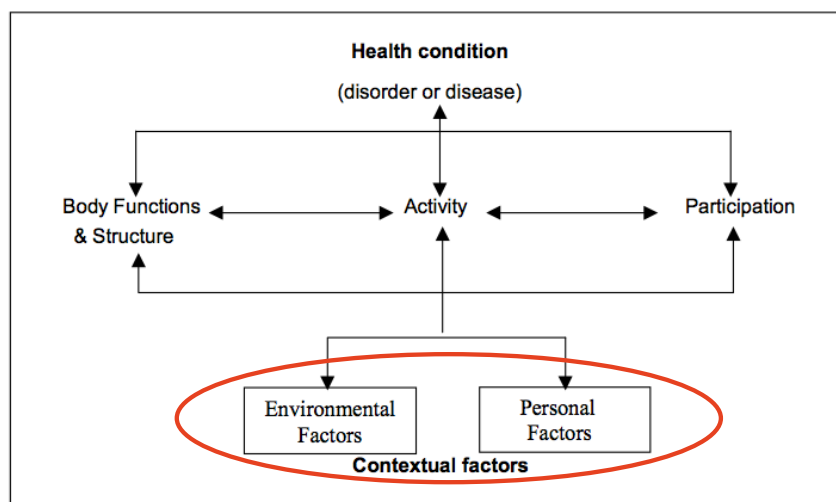
### Our experience:

- Life book/Memory book aids transition to residential care: who the person is, what is important to them

## Interventions targeting contextual factors

The University of Sydney

Page 115



The University of Sydney

Page 116

## Behaviour modification: Intervention for Behavioural and Psychological Symptoms of Dementia (BPSD)

- Clare O'Connor: Tailored activities program
- Phillip Lee: Management of behavioural symptoms in FTD in the clinical setting (9<sup>th</sup> ICFTD Caregiver Conference, 2014)
- The IPA Complete Guides to *BPSD* – Specialists Guide (2015)

<https://www.ipa-online.org/publications/guides-to-bpsd>

The University of Sydney

Page 117

## Psychosocial support

### For person with PPA

- Referral to Alzheimers Association, Association for FTD
- Refer to psychological support services (GP Care Plan for mental health in Australia)

### For partner

- Respite
- Social support
- Refer to psychological support services (GP Care Plan in Australia), grief and adjustment counselling
- Problem-based coping interventions (Mioshi & colleagues)

The University of Sydney

Page 118

## Target lack of information with education



### Communication and Support group

- Monthly meeting at War Memorial Hospital for individuals with PPA and family members 8-10 per group is ideal
- Peer matching on severity to avoid confrontation with greater impairment
- **Information:** low % know what aphasia is, seek information about what PPA is, what is happening to language abilities. 1 hour session with neurologist
- **Psychosocial support:** peer support, empowers people in their expertise to help each other
- Exchange coping, problem solving, communication, relaxation strategies (Sapolsky et al., 2011)

The University of Sydney

<http://upload.wikimedia.org/wikipedia/commons/5/5c/East0083.jpg>  
Accessed 15Jun11

## Target lack of information with education



### Clinician mediate access to educational resources

- Highlight need for caution when accessing information on internet
- fear and depression after internet browsing
- E.g. tips from websites of pwPPA

### PPAmphlet (PPA pamphlet)

- Quarterly newsletter (Cathy Taylor)
- e.g. piece by Michael J Fox on attitude
- Card explaining PPA for drivers

The University of Sydney

<http://upload.wikimedia.org/wikipedia/commons/5/5c/East0083.jpg>  
Accessed 15Jun11





**AAA (Australian Aphasia Association) Conference 2010  
included a half-day session on PPA**

Programme	
An update on Primary Progressive Aphasia	Karen Croot
My Life in the Theatre & PPA	Man with PPA
A Conversation about Living with Primary Progressive Aphasia	Daughter of woman with PPA & Cathy Taylor
Living with Primary Progressive Aphasia	Wife of man with PPA

The University of Sydney

Page 121

## Target lack of information with research

- Many pwPPA and their families are willing to participate in research if invited
- A means of accessing further information or experimental treatments, or to create meaning/positive benefit for others out of their adverse experience
- Some do not want the additional burden of research appointments, confrontation with disease

**All researchers here at the International Conference on the Frontotemporal Dementias**

**The German Context:**

- **FTD-Netz:** Prof Janine Diehl-Schmid, Munich  
<http://morbuspick.blogspot.de/p/ftd-netzwerk.html>
- **DZNE:** Peter Nestor, Magdeburg recruiting pwPPA, prospective study, can refund travel costs [peter.nestor@dzne.de](mailto:peter.nestor@dzne.de)

The University of Sydney

Page 122

## Raising awareness: Consumer voices

- **Stories of pw PPA:** Joann Douglas, Agnes Summers & Jade Cartwright, Laurie Sacks <http://looklikelaury.com/about-the-film/>
- **Carer stories:** Shirley Rutherford
- **Creative works**
  - Sculpture/Installation with poem by Mrs CB
  - Play about PPA: **Water Falling Down**
  - Art by Ann and Sophie Cape

### June Andrews, University of Stirling

- Avoid representations that amplify community fears and stigma
- Helpful to provide sensible information in plain language

<https://theconversation.com/still-alice-is-far-from-a-good-thing-for-dementia-awareness-38007>

The University of Sydney

Page 123



Agnes Summers  
(top) and Jade  
Cartwright

## Shirley Rutherford

*Aphasiology*, 2014  
Vol. 28, Nos. 8-9, 900-908, <http://dx.doi.org/10.1080/02687038.2014.930812>



### Our journey with primary progressive aphasia

Shirley Rutherford

**Background:** My husband Boyd and I had a 6-year journey with primary progressive aphasia (PPA) that held many challenges for us along the way. This article describes that journey.

**Aims & Main Contribution:** I hope that hearing about our experience may be helpful to other people with a family member who has PPA, and provide clinicians and researchers insight into the PPA journey.

**Conclusions:** I hope that through more research, there can be more understanding about PPA and consequently more support for families with members suffering this cruel disease.

**Keywords:** Nonfluent progressive aphasia; Caregiver; Personal story; Lightwriter; Communication book; AAC.

This is our journey with this awful disease. I hope it might be helpful to other people going through this disease with a family member to hear about our experience. I did my best to keep everything as normal as possible for as long as I could. Now that our journey with PPA is written down, I feel a lot better. It is as if a heavy load as been lifted off my shoulders. I know I can now start moving forward. It will take time but I need to do my best to make a life for myself. That's what Boyd would have wanted me to do. I just hope that more research can be done. The more understanding about PPA there is, the more support there will be for families with members suffering this cruel disease.

The

Page 124

## Mark Swivel

### Note on Aphasia

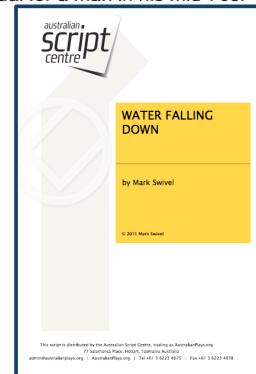
The character of Dad has a condition known as primary progressive aphasia, a form of dementia that, among other things, attacks or corrupts language. Its causes are mysterious and its effects fitful, attended by the odd moment of fluency during a slow decline into senility and physical incapacity. The primary expression of the condition in Dad's case is a loss of nouns. Hence, Dad often searches for the words for 'things' throughout the play. It is important to state that Dad knows the words he is searching for but cannot articulate them (although occasionally a word does 'come'). The Son fills in the gaps in Dad's speech caused by the aphasia. Dad does not have Alzheimer's disease or any other damage to his memory other than is usual for a man in his mid-70s.



© 2011 Mark Swivel  
The University of Sydney

<http://www.stagewhispers.com.au/reviews/water-falling-down>  
Accessed 16Jan13

Page 1 of 83



## Ann and Sophie Cape



<http://www.dailytelegraph.com.au/newslocal/north-shore/mosman-artists-ann-and-sophie-cape-create-exhibition-focusing-on-the-personal-pain-of-dementia/news-story/0c9667cdf9b0e6337233893b3918089a>

Accessed 22Aug16

## Raising community awareness

- Educate clinical students – changing the future
- Educate primary health care physicians and allied health care providers  
E.g. NSW Dept of Health Agency for Clinical Innovation
- Cultural events: Film nights, art shows (Frontier Sydney)
- Media: Cartwright & Elliot on ABC television



The University of Sydney

## Community access

- Safely Home programme: Collaboration between Alzheimer's Australia and State Police Forces
- <https://nsw.fightdementia.org.au/nsw/support-and-services/services-and-programs-we-provide/safely-home>
- Register photo and details with Police
- Nominal Fee

ALZHEIMER'S IMPEDES  
AUSTRALIA EMPOWERS  
FIGHTDEMENTIA.ORG.AU

### Ideas to encourage someone to wear their Safely Home Bracelet



- 1. Wrap the bracelet in a box and give as a present**  
You could give the gift on the person's birthday, Easter or just a reason to be kind.
- 2. Have a grandchild or other loved one present the bracelet**  
The person with dementia may appreciate the gesture, cherish the bracelet, and wear the bracelet even if not initially taken with the style.
- 3. Ask your GP to give the Safely Home bracelet**  
If the person with dementia has a doctor's appointment soon after receiving the bracelet, ask the GP to place the bracelet on the person during the appointment. The person with dementia may better receive it.
- 4. Place the bracelet on the person's dominant hand**  
This will make it more difficult for the person to try to release the clasp and remove the bracelet. Although the clasp does need to two hands to release it.

The University of Sydney

Page 128

## ANT's community access 2010-2016

### Speech Pathologist

- Provided speech and language assessment, education and interventions, Communication and Support Group
- Supervised intern/student speech pathologists working with ANT
- Referred ANT for research projects
- Encouraged PoA and Guardianship
- Arranged
  - Communication cards and life book
  - Introduction card, Driving card, YES/NO cards
  - Communication cards for hospital
    - I have pain (with rating scale)
    - Can I see the doctor?
    - I need to go to the bathroom
  - Computer training course offered by local council
  - Communication partner visits from local church
  - Attendance at AAA Conferences
  - AAC program on i-pad, telephone relay service (TTS)
- Visited bank to assist with change to PIN numbers

The University of Sydney

Page 129

## ANT's Introduction Card

I have a rare medical condition

**Primary Progressive Aphasia**

I have difficulty speaking but can understand everything you say

**Please be patient with me**

The University of Sydney

Page 130

## ANT's community access 2010-2016

### Friends

- Power of attorney, telephones and talks to ANT
- Social participation: dinners, holidays

### Neighbour

- assists with tradesmen & home maintenance, transport to speech therapy, communication with health professionals

### Community organisations

- Local stroke/aphasia group
- Friday Seminar at Church
- Tai Chi

### ANT herself: **extremely proactive in finding services and activities and asking for assistance with access if required**

- Preferred method of communication was email

The University of Sydney

Page 131

## Resources

For a full reference list from this workshop, and other resources for assessment and intervention in PPA, please visit our website at the

**ARC Centre of Excellence in Cognition and its Disorders (CCD)**

**<https://www.ccd.edu.au/>**



**Currently under construction:**

**Please visit from October 2016 onwards!**

The University of Sydney

Page 132

## Thank you



**Professor Diehl Schmidt and the organisers of 10th  
International Conference on Frontotemporal Dementias  
August 31-September 2, 2016 Munich/Germany**



The University of Sydney

Page 133

## Thank you

**ANT, TOC, WIC and their spouses/families/neighbours**

**Maya Henry**

**John Hodges**

**Cristian Leyton**

**Wendy Longley**

**Rosemary Manusu**

**Lyndsey Nickels**

**Leanne Ruggero**

**Emily Rogalski**

The University of Sydney

134

Page 134





## Our support

**Alzheimer's Australia Research**  
Hazel Hawke Research Grant in Dementia Care

**Australian Research Council**  
Centre of Excellence in Cognition and its Disorders

**Jackstaedt Stiftung**  
P.D. Dr Steffie Abel (Aachen, now Manchester)

**Robert Bosch Stiftung**  
Ms Theresa Raiser (LMU, Munich)

**Australian Government Endeavour Fellowship**  
Ms Inga Hameister (RWTH Aachen, now Macquarie University Australia)

The University of Sydney







135
Page 135



<http://www.dailytelegraph.com.au/newslocal/north-shore/mosman-artists-ann-and-sophie-cape-create-exhibition-focusing-on-the-personal-pain-of-dementia/news-story/0c9667cdf9b0e6337233893b3918089a>

Accessed 22Aug16